

Cystic fibrosis in Russia: background and a model for future collaboration with the West

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HISTORICAL

A search of medical records identifies the first cases of cystic fibrosis (CF) identified in Russia in 1967. It was not until 1989 that two officially designated CF centres opened, the first one in Moscow and the second one in St Petersburg. Within the subsequent 5 years a further six centres were recognized in other cities.

Over the last 5 years there have been huge political changes with resulting changes in geographical demarcation. The former USSR has broken up into a number of republics and some of these, such as the Ukraine and Byelorussia, have developed recognized CF centres of their own. Russia itself has a population of approximately 150 million.

INCIDENCE

In European Russia careful calculations have shown there to be approximately one CF child in every 5500 newborns. This would equate with approximately 650 new cases every year. Moscow itself would produce approximately 30 children per year with CF.

IDENTIFICATION

There is no screening programme at present in Russia and it is recognized that most cases are undiagnosed. Even doctors training as paediatricians may never have seen a case of CF and there is clearly a major need for raising the profile and awareness of the condition as well as improving diagnostic services. Accurate sweat testing may be available in some of the larger cities in Russia but access to these facilities is very difficult and still depends on the clinical acumen of the local doctor recognizing this diagnostic possibility.

GENETIC PROFILE

For interesting political and historic reasons genetic research has had a very high profile in Russia. There is good international collaboration and genetic studies of large CF populations have been undertaken. In one study of 230 CF

patients, aged between 3 months and 28 years, the Delta F508 deletion was present in 52%. All these patients were pancreatic insufficient. The study may show a bias because children with pancreatic sufficiency may be under-represented. Further studies are currently under way.

CLINICAL STUDIES OF CYSTIC FIBROSIS IN RUSSIA

In spite of poor facilities and inadequate funding, a small team of dedicated doctors have attempted to review and describe the clinical features of CF. Over a 10-year period a study of 186 CF patients was undertaken to identify the most important prognostic features. During the period of study there were 41 deaths in the first year of life and of these 20 (50%) had severe pneumonia or lung abscess.

The incidence of *Pseudomonas* has been steadily increasing (this may represent better ascertainment) and in the current clinic population approximately 70% of children have *Pseudomonas*. No cases of *Pseudomonas cepacia* have ever been found. Sixty-one per cent of children were also found to have *Staphylococcus aureus* and 45% have *Haemophilus influenzae* isolated from the sputum.

Chronic liver disease, clearly identifiable as biliary cirrhosis, was found in 13% of the patients. Four of the cases developed massive haematemesis and three of these died. Fifty-two per cent have developed some abnormality of glucose intolerance although none have developed frank diabetes.

The range of antibiotics available in the West are only sporadically available in Russia but in spite of this some trials have been carried out with aminoglycosides and with third generation cephalosporins or quinolones.

PHYSICAL THERAPY

For many years there has been the development of some special Russian forms of physical therapy including decimeter and sinusoidal modulated currents and different forms of mechanical vibration. Also some research studies have looked at the regular use of treadmill, veloergometer and trampoline.

More conventional physiotherapy was also part of the clinic practice. Children had breathing exercises, simple

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percussion and exercise and these were recommended twice a day although compliance was variable.

110 CF children were able to take part in a period of treatment in a sanatorium in the southern part of Crimea where there was less air pollution and a daily programme of professional input. Unfortunately this type of treatment is at present only available for CF children with mild or moderate disease. There are also major funding problems in the development of CF holidays and camps.

PROGNOSIS FOR CF IN RUSSIA

Until very recently the prognosis in identified cases has been very poor with an average life expectancy of around 14 years.

PROFESSIONAL SERVICES AVAILABLE FOR CF IN RUSSIA

The whole of the management structure for CF care is based on the doctor as the only professional. There are no routine services available from physiotherapists, dietitians, specialist nurses, social workers and others that one would normally find in the CF team in the West.

Students decide on entering medical school whether they will go through an adult training or paediatric training and thus the paediatricians who currently look after CF have only studied in a paediatric medical school. On qualification they go through a programme of internship which may include specialist training to include nutrition and dietetics and physiotherapy. The doctors in a CF team will then have the role of a doctor as well as one of the other paramedical professions.

The CF team in Russia would thus have more doctors on the staff than an equivalent sized clinic in the UK but the support staff would be completely lacking. Doctors themselves would have to do the physiotherapy on inpatients and also undertake the nutritional calculations and have a central role in discussing diets.

The role of nurses is also somewhat different and there are not at present equivalents within the Russian setting of clinical nurse specialists or nurse practitioners.

When children are not in hospital their medical care is undertaken by the local polyclinic where paediatricians will be dealing with both acute and community type problems but may have very little or no knowledge of CF. Communication between the hospital clinic and the polyclinic doctors may be variable and only rarely would parallel close shared care as one might get in the West.

This model of health care is totally different from the Western model and has to be clearly recognized if good collaboration with the West is going to take place.

Other Support for CF Children

Parents' groups for CF have been developed for some time and are now increasingly developing in parallel ways to the Western model.

The president of the Russian Cystic Fibrosis Association is actively involved in obtaining charitable support, help and supplies of essential medicines not only for the large city units but right across Russia.

Complementary Medicine

Russia has a rich history in the development of folk medicine. Many Russians are firm believers in traditional remedies and some combine this with a distrust of modern medical care. Within the CF clinic there is considerable use of traditional remedies as well as modern commercially promoted tonics, elixirs, herbs and vitamin supplements and remedies that are claimed to improve bile flow, etc. A few children have also been given urine therapy. Some of these are expensive and there is no good evidence showing that they have a particular merit in CF.

Parental Perceptions

Parents of newly diagnosed CF children are truthfully told that with existing resources prognosis is poor and their children may not live beyond their teens. Most Russian families only have one child and in most families both parents work.

One parent, usually the mother, may stop working to look after the child, knowing that he/she may die. When their child becomes ill he/she is usually admitted to hospital and it is somewhat arbitrary whether there will be appropriate antibiotics for full and effective treatment.

Before the development of good social support a family with only one child who has CF, knowing the poor prognosis and with a reduced income, could easily be in despair. It may even make their despair worse when they hear that in the West children with CF can now expect a fairly normal life, with attendance at school, and may well grow up into adult life and train professionally, marry and even become pregnant. Their doctors may also share this despair with increasing recognition that they could do so much better if only the resources were available.

It is in this setting that a Winchester based charity, International Integrated Health Association (IIHA), started a programme which is presented here as one model of good collaboration.

THE SOUTHAMPTON MOSCOW TWINNING: A MODEL FOR COLLABORATION

For many years it has been possible, occasionally, for doctors in Russia to attend conferences in the West with sponsorship from pharmaceutical companies or international charities

and agencies. These visits are exciting and interesting but often give little insight into the actual ways in which healthcare is delivered in the host country. It was also not infrequent to have visits from overseas professionals or to have contacts from representatives of pharmaceutical companies who might bear gifts of some of their own products to be tried. These forms of contact, although welcome, do not fundamentally change the nature of health care and there is a risk that the givers of charitable gifts might have a slightly patronizing attitude towards their colleagues and the receiver of these gifts may develop a 'charitable mode' where gifts are simply received and consumed and then further gifts and donations are requested.

In 1993 representatives of the International Integrated Health Association made contact in Moscow with both the Head of the Cystic Fibrosis Service and the President of the Cystic Fibrosis Parents Association. That visit was followed by a professional fact-finding visit from the Director of the Southampton Cystic Fibrosis Service and as a result a planned programme, over a period of 2 years, was established with a set of clear objectives:

- (1) To make available a constant supply of modern microsphere enzymes. This supply was not to be given simply as a charitable gift but was in the setting of a clinical trial which would provide useful and possibly unique data. It would also be used as a lever for developing a permanent supply of enzymes in the long term, not only in Moscow but throughout Russia.
- (2) To have a regular close exchange of medical and paramedical staff between the two units with a quite specific intention of an exchange of ideas and clinical practice at patient care level.
- (3) To develop a series of research projects which would facilitate the entry of the Moscow clinic staff into the international world of pharmaceutical trials, collaborative projects, international conferences and exchanges. It was recognized that for historic reasons the Russian medical fraternity did not have a high research profile and had an uncertain level of credibility. In order for suspicion in the eyes of grant giving bodies to be allayed, it was important that the clinic produce protocols and research of the highest order. Part of the programme was to make the clinic staff feel self-sufficient with an international standing in their own right. It was recognized that both sides of the exchange could learn from the other and that a good collaboration would be symbiotic. For example, it was certainly easier for the Southampton clinic to obtain a grant and shoulder the 'credibility issue'. On the other hand, the Moscow team had a large population of children who were not getting optimal care but whose parents were very willing to

take part in any trial and whose doctors were very keen to participate.

- (4) To undertake a review of patient care plans and to develop, where possible, an optimal routine outpatient based service rather than the present crisis management. This would include the use of Western style hospital records, growth charts and communication network, with the polyclinics. There would also be a parallel programme of parent support and self-help,

Two Years of Twinning

Year One

The major research study in the first year was to assess the effect of providing optimal modern enzymes and nutritional support while leaving the rest of management as it had been previously. This study was made possible because of the combined efforts of the International Integrated Health Association and a major grant from Solvay Duphar. It was possible to provide this single change in management in half of the children (approximately 45) who had full care in the clinic. The detailed results of this study are presented elsewhere. In summary, it was shown that with this nutritional support alone the children improved dramatically in their growth and wellbeing and there was a reduction in the amount of respiratory disease.

A number of exchanges were undertaken including regular visits by groups of the CF team from Moscow to Southampton, the joint attendances at conferences and visits by Southampton personnel to Moscow. The British visitors included two physiotherapists, a dietitian, a microbiologist, a clinical nurse specialist and even a medical student undertaking a project.

At the same time that these clinical contacts were made IIHA was arranging visits of parents and patients and also made major steps in developing the concepts of self-help and support at the local level.

Year Two

There was a continuation of the nutritional support for the first group of children and the other full care children were also recruited in but given vitamin supplements and a more structured clinic environment. The analysis of the second year is not yet complete.

The results of the first year studies have been presented by both the Russian and Southampton teams at two international conferences, three national conferences in Russia and one national conference in the UK.

A clinical service is now well established in Moscow with regular 3 monthly outpatient visits, Western style documentation and enthusiastic parents' support group.

The Future

With the increased confidence and credibility of the Moscow team there is now the rapid development of a networking within Russia with the Moscow service as the resource centre and funding has already been promised for Russian CF conferences and clinical visits at an individual level.

There is enthusiasm to maintain continuing collaboration. Plans are already underway to extend twinning between

Southampton and Moscow in other clinical departments such as immunology, asthma and diabetes.

The twinning between Southampton and Moscow has been an unqualified success for patients, for doctors and for research collaboration. It is strongly recommended that twinning between other centres in the West and in Russia be undertaken, in view of the benefits in both directions.